PRISM SECTION

(Patients' Revelations as Insightful Studies of their Management)

HENOCH–SCHÖNLEIN PURPURA PRESENTING AS ABDOMINAL PAIN BEFORE PURPURA: A CASE REPORT

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INTRODUCTION

Henoch–Schönlein purpura (HSP) is an acute, systemic, immunoglobulin-mediated small-vessel vasculitis. It is the commonest vasculitis of childhood and is typically characterised by a tetrad of abdominal pain, arthritis, palpable purpura, and renal disease. All patients develop palpable purpura, while 84-90% develop arthritis,[1,2] 57-58% develop abdominal pain,[1,3] and 20-54% develop renal involvement.[1,2,4] Gastrointestinal symptoms can be the first presenting complaint with the absence of initial purpura, leading to a delay in diagnosis. A case that presented this way is reported.

CASE REPORT

A 3-year-10-month-old girl first presented to a paediatric emergency department with colicky abdominal pain and constipation for three days. She was known to have chronic symptoms of constipation, passing hard stools once every two to three days, associated with straining. There was no per rectal bleeding, no vomiting, or symptoms of recent viral illness (including upper respiratory tract symptoms and fever). She was otherwise clinically well. The clinical impression was constipation colic.

Six days after the onset of symptoms, she presented a second time to the emergency department because the abdominal pain persisted. She was clinically well. The decision was made for referral to the paediatric specialist outpatient clinic with the provisional diagnosis of constipation colic.

Ten days after the onset of symptoms, when reviewed at the specialist outpatient clinic, the abdominal pain had resolved, but she was noted to have a left ankle bruise. The parents also reported that the child was reluctant to walk, complaining of left ankle pain since that morning. There was no recent history of trauma or easy bruising.

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On examination, the child was well and there was no abdominal tenderness or palpable masses. There were three small (1-2mm) papules bilaterally on the lower legs and also a 3cm bruise (non-blanchable but not palpable) over her left medial ankle. No visible swelling of the ankle was noted, the range of motion was full, and her gait was normal. Xerotic skin on both heels and shins were noted.

The working diagnosis at the paediatric specialist clinic was constipation colic, with possible eczema in view of the dry skin. Dietary advice was given to the mother, lactulose prescribed for constipation, and topical hydrocortisone for the skin lesions. As the bruise was not typical of eczema, an early follow up was given to review this.

Twelve days after the onset of symptoms, the patient presented again to the emergency department for the third time, as there were now multiple palpable bruises on both the lower limbs. Abdominal pain had also recurred. She was less active and her blood pressure was noted to be 122/85 mmHg, which was high for her age. The normal systolic pressure for a child aged three to four years ranges from 86-112 mmHg.

The clinical diagnosis of HSP was then made, and the patient was admitted for further work up. In the ward, her blood pressure improved to 87/52 mmHg.

Laboratory tests (shown in Table 1) consistent with the diagnosis of HSP included a normal platelet count. There was gut wall oedema visible on ultrasound. Urine microscopy and urine protein:creatinine ratio were normal.

The patient was started on prednisolone 15mg per day (1mg/kg/day) and given symptomatic treatment with paracetamol, ranitidine, and brufen. The abdominal pain resolved and she was discharged 36 hours after admission. She was given an early follow-up appointment in one week, with urine dipstick and urine protein:creatinine ratio to exclude renal involvement. The follow-up test results were normal. Prednisolone was continued for two weeks at 15mg per day, then tailed down to 7.5mg per day for one more week.

DISCUSSION

Absence of palpable purpura

Diagnosing atypical HSP without a rash was the challenge in this patient. The appearance of bilateral palpable purpura on both lower limbs on day 12 clinched the diagnosis. The patient had no renal complications and the symptoms of joint pains rapidly settled with prednisolone. On follow-up, she showed continued resolution of the joint pains and the rash had resolved.

Table 1. Laboratory investigations done on admission to the paediatric inpatient ward
Haematology Total white count: 13.09 X 10^9/L (Normal value: 5-15 X 10^9/L) Differential count: Neutrophils 61.9%, lymphocytes 23.1%, eosinophils 1.0% ESR: 16mm/hr (Normal reference range: 3-9mm/hr) Serum creatinine: 17μmol/L (Normal value: 28-55μmol/L)
Urine Microscopy per high power field: Red blood cells 1, white blood cells 2, epithelial cells 1 (Normal values: Red blood cells 0-2, white blood cells 0-4, epithelial cells 0-5) Urine protein to creatinine ratio: 0.02g/mmol (Normal value: <0.02g/mmol)
Abdomen ultrasound No intussusception. Thickened bowel wall was noted in the gut at the epigastrium, left hypochondrium, and left iliac fossa regions.

Henoch-Schönlein purpura with no rash

The presentation of HSP atypically without an initial rash is reported in the literature.[5,6] In a child with only abdominal pain, the differential diagnosis of abdominal pain should also be considered — intussusception, appendicitis, incarcerated inguinal hernias, peptic ulcer disease, gastroenteritis, urinary tract infection and constipation. A high index of suspicion is necessary in a child with persistent abdominal pain, and an abdominal ultrasonography can support the diagnosis of HSP.[5] Careful skin examination is also important as this was what led to the suspicion of HSP in this case.

Pathophysiology

In HSP, IgA-containing immune complexes deposit in small vessels predominantly in the dermal, gastrointestinal and glomerular capillaries. The resultant inflammatory response and small-vessel vasculitis cause the skin manifestations of petechiae and palpable purpura. In the intestinal wall, the vasculitis may manifest as gastrointestinal bleeding and intussusception. In the glomeruli, it can manifest as haematuria, proteinuria or nephropathy.

Pathophysiology

It is useful to review the usual presentations of HSP. Ninety percent of cases occur in childhood.[7,8] It usually occurs between 3 and 15 years, with a mean age of onset between 6 and 7 years of age.[9]

In a prospective study of 223 newly diagnosed HSP cases in Finland, the initial presenting complaint was purpura in 73%, arthritis in 15%, and gastrointestinal symptoms in 11% of patients.[1] A retrospective Italian review yielded similar results, with purpura, arthritis and abdominal pain as the presenting complaint in 74%, 15% and 12% of the patients respectively.[2] HSP may also present with scrotal pain and swelling.

Complications of HSP include fulminant purpura with blisters, and, less commonly, neurologic manifestations such as headaches and seizures. There may be a precipitating viral or bacterial illness.

Typically, HSP presents with a tetrad of purpura, abdominal pain, arthritis or arthralgia, and renal disease. The symptoms and signs can develop over days to weeks, and may vary in the order of presentation:

1. ***Palpable purpura in the absence of thrombocytopenia or coagulopathy.*** The rash usually begins with erythematous, urticarial and macular wheals. It then coalesces and develops into ecchymoses, petechiae, and palpable purpura. It often manifests in a symmetrical pattern at pressure-dependent areas. In non-ambulatory children, the face, trunk, and upper extremities may be more affected.[7]

2. ***Abdominal pain.*** Gastrointestinal manifestations such as nausea, vomiting, abdominal pain, transient paralytic ileus, gastrointestinal bleeding and intussusception can occur. In a retrospective study conducted in Taiwan on 261 children with HSP, 58% developed abdominal pain.[3] Gastrointestinal symptoms usually develop within 8 days of the appearance of the rash, but can also be found to precede skin manifestations in 15–35% of cases.

3. *Arthritis or arthralgia.* Arthralgia occurs in 84-90% of HSP patients.[1,2] Large joints of the lower extremities are most commonly affected. This is usually transient or migratory, and typically oligoarticular (1-4 joints).

4. *Renal disease.* Renal involvement occurs in 20–54% of patients.[1,2,4] Haematuria and/or proteinuria (without renal function and blood pressure abnormalities) to acute nephropathy with renal impairment may occur.

Investigations to confirm HSP
When HSP is suspected, a full blood count may show normochromic anaemia if there is gastrointestinal bleeding; leukocytosis and raised erythrocyte sedimentation rate if there was a preceding bacterial illness; and platelet and coagulation profile should be normal (to rule out thrombocytopenia or coagulopathies resulting in a similar purpuric rash).

Urinalysis should be done to look for haematuria or proteinuria, which can occur in renal involvement. If urinalysis is abnormal, a renal panel should be done to monitor the creatinine level. There is no laboratory test diagnostic for HSP, but serum IgA can be high in 50-70% of cases.[10]

Management of HSP
Management is mainly symptomatic and supportive, and can be done in the outpatient setting if the patient does not require admission. Ensure that the hydration status is adequate. Symptomatic treatment with paracetamol or NSAIDs may be prescribed for pain and joint swelling. The use of oral corticosteroids is still controversial, but some studies have shown that it can shorten the duration of abdominal pain. If used, oral prednisolone 1-2 mg/kg/day for 7 days is recommended.

Admission should be considered if the following features are present: [7]
- Inability to maintain adequate hydration with oral intake;
- Change in mental status;
- Severe joint involvement limiting ambulation;
- Severe abdominal pain or gastrointestinal bleeding; or
- Renal insufficiency (elevated creatinine), hypertension, or nephrotic syndrome.

Complications of HSP
Gastrointestinal bleeding, bowel ischaemia or necrosis, intussusception and bowel perforation may occur. An ultrasound of the abdomen should be ordered if intussusception is suspected.

Renal manifestations usually occur in the first 28 days after the initial presentation. Gross or microscopic haematuria, proteinuria or nephritic syndrome may be present.[4] In a 2005 systematic review, Narchi followed up children with HSP and found that 1.8% of the total study population developed subsequent renal impairment.[11]

Follow up for renal abnormalities and blood pressure
Blood pressure and urinalysis should be monitored for up to 6 months after the initial presentation.

CONCLUSION

- The key take-home message in this case report is that HSP can present atypically with just persistent abdominal pain for days before appearance of the characteristic rash.
- A high index of suspicion for HSP when dealing with a child presenting with abdominal pain and joint pain is needed. Follow up for the appearance of palpable purpura and advice to the caregiver on what to expect will help in earlier diagnosis.

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